

BACKGROUND

- UTROSCT is a rare mesenchymal uterine neoplasm composed of more than 50% sex-cord-like cells.
- A benign and well circumscribed tumour, however there is a low malignant potential with a **5.9%** recurrence rate.
- Clinical symptoms of UTROSCT are non-specific.
- True myometrial invasion is uncommon but can be present necessitating a more radical cytoreductive approach.

Factors associated with aggressive behaviour:

- >2 mitoses per 10HPF
- Extrauterine spread on imaging
- GREB1 rearrangement
- Presence of necrosis

CASE HISTORY

- 45-year-old Afro-Caribbean female presented with chronic abdominal pain.
- US identified a heterogeneous myometrial mass which was characterized with MR and CT, showing a **16cm uterine mass with features suspicious for sarcoma**.
- At laparotomy a 35cm uterine mass was noted, adherent to both ovaries, small bowel, bladder and rectosigmoid. A total hysterectomy and bilateral salpingo-oophorectomy with small bowel resection and side-to-side anastomosis were performed to achieve complete cytoreduction. The patient had an uneventful recovery.
- Histological analysis showed a myometrial tumour (**Fig 1**) with a microfollicular growth pattern, vesicular nuclei and occasional nuclear grooves. Tumour with the same microarchitecture was lining a left ovarian cyst (**Fig 2**) → **FIGO Stage 2A** pathological staging.

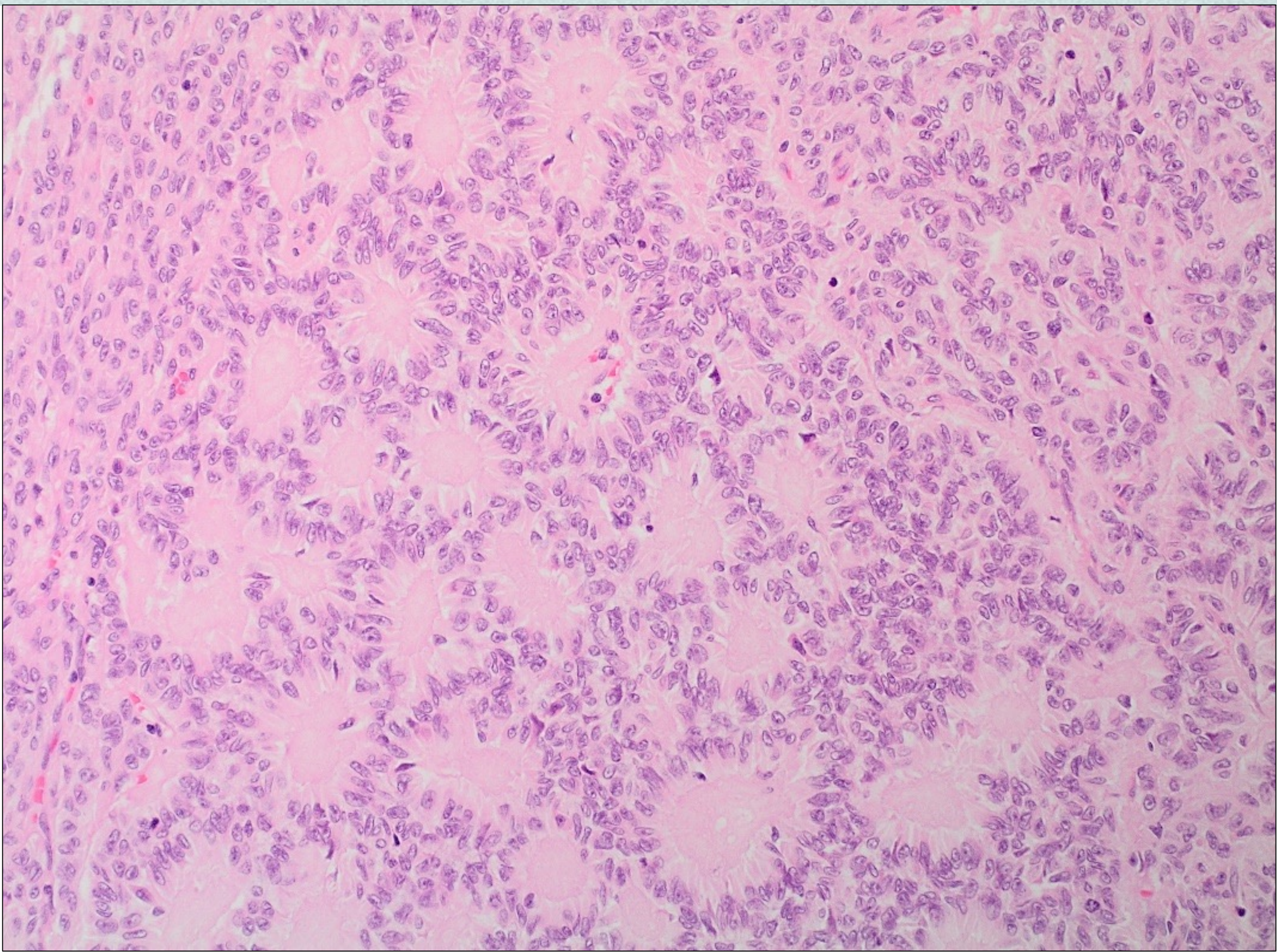


Fig 1. Myometrial mass HE x 200: Tumour with microfollicular pattern; cells showing vesicular nuclei, occasional nuclear grooves, and pale cytoplasm.

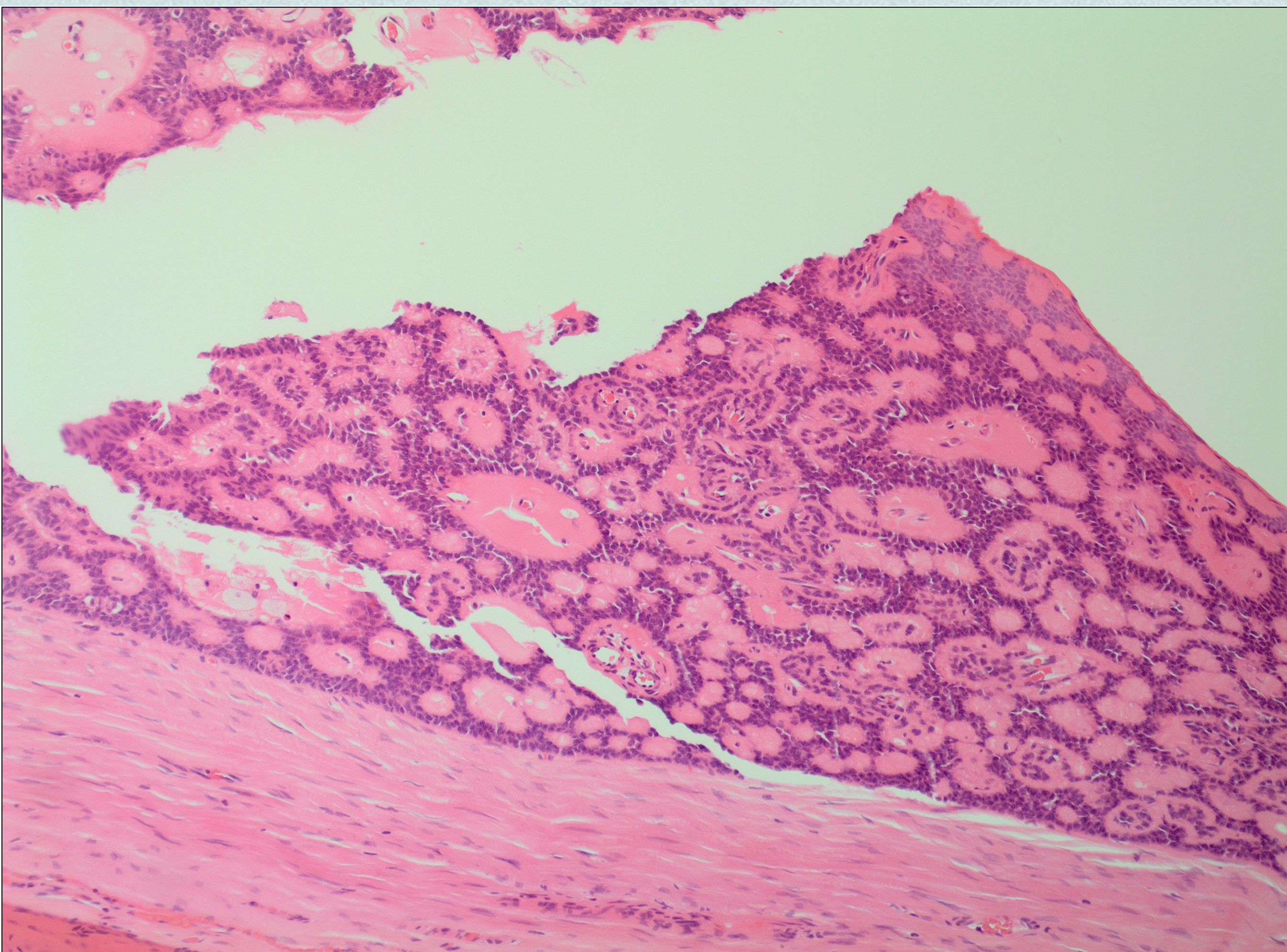


Fig 2. Ovarian mass HE x 100: Tumour nodules, similar to the myometrial mass, lining a left ovarian cyst.

DISCUSSION

- Histological diagnosis is challenging as these tumours co-express epithelial, smooth muscle, sex-cord markers, as well as steroid receptors.
- Due to ovarian involvement, the main differential diagnosis in this reported case was an ovarian sex-cord stromal tumour metastasizing to the myometrium.
- FOXL2 alterations seen in adult granulosa cell tumours and DICER1 alterations seen in Sertoli-Leydig cell tumours were both negative and were valuable in reaching the diagnosis.

CONCLUSION

UTROSCT is a challenging histological diagnosis that impacts on patient counselling, management and outcome.

Due to paucity of evidence, individualised follow-up protocols are devised based on specific risk factors for recurrence and aggressive behaviour. UTROSCT cases should be managed by specialized gynae-oncology teams in cancer centres.

Umeda S; Tateno M; Miyagi E; Sakurai K; Tanaka R; Tateishi Y et al. (2014) Uterine tumors resembling ovarian sex cord tumors (UTROSCT) with metastasis: Clinicopathological Study of two cases. International journal of clinical and experimental pathology. 7(3):1051-9.
Goebel EA; Hernandez Bonilla S; Dong F; Dickson BC; Hoang LN; Hardisson D et al. (2020) Uterine tumor resembling ovarian sex cord tumor (UTROSCT): A morphologic and molecular study of 26 cases confirms recurrent NCOA1-3 rearrangement, The American Journal of Surgical Pathology. 44(1), 30-42.
Chang, B. et al. (2020) Recurrent uterine tumors resembling ovarian sex-cord tumors with the growth regulation by estrogen in breast cancer 1-nuclear receptor coactivator 2 fusion gene: A case report and literature review. Diagnostic pathology. 15(1), 110.
Marrucci, O. et al. (2019) Uterine tumor resembling ovarian sex cord tumors type II with vaginal vault recurrence, Case Reports in Obstetrics and Gynecology.